ATHETOSIS, ITS TREATMENT AND PATHOLOGY.*

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SINCE the time when athetosis was first described by Dr. W. A. Hammond, in 1871, a large number of cases have been reported both in this country and in Europe.

The subject of spasm, both fixed and mobile, has been studied very thoroughly within the last few years, and the result has been to classify all localized spasmodic diseases due to cerebral lesions, as exemplified by athetosis, chorea, ataxia, tremor, and tonic spasm, as allied affections, all depending upon lesions occurring in the same projection-system. Dr. Hammond, in his original article, located the disease in the optic thalamus or corpus striatum. Oulmont, in his monograph on athetosis, attributes the disease to what he calls "athetoic fibres," which he supposes to exist in the posterior part of the internal capsule in front and outside of the sensory tract. Denange states that all forms of spasm have a common origin, and that they may be produced by a lesion in any part of the motor tract. In this view, Sharkey fully concurs, and I believe that opinion is generally held at the present time. That lesions of a certain part of the motor-tract will produce mobile spasms, is undoubtedly true, but that lesions of any part of the motor tract will produce tremor, ataxia, or athetosis, is not, to

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my mind, supported by fact. I shall endeavor to show in this paper that athetosis and its allied disorders are seldom the result of lesions in the motor tract, and that when the lesion does occur in the motor tract it is confined to a particular part of it.

Athetosis is usually, not invariably, preceded by hemiplegia or epileptic or epileptiform siezures. Cases have been reported by J. Coates, George Ross, Fletcher Beach, Landouzey, and others, in which the patients had neither suffered from paralysis nor epilepsy in any form. When there has been hemiplegia, the athetosis does not usually make its appearance for a considerable length of time. Sometimes several years elapse between the advent of the hemiplegia and the appearance of the athetosis.

In a large proportion of cases in which the athetosis has been preceded by hemiplegia, it is a noticeable fact, and one well worthy of attention, that either the paralysis disappeared before the athetosis became apparent, or else the athetoïc movements increased in direct proportion as the hemiplegia decreased. I think that this fact is of pathological interest, as I shall attempt to show later.

On the treatment of athetosis there is very little to be said. The very nature of the lesions which have been found to produce athetosis, precludes the possibility of their ever being removed by any remedial measures that we are able to resort to at the present time. But if we are unable to remove the cause of the disease, I think the following cases tend to show that the manifestation of the disease can be arrested for a longer or shorter period of time, and in some cases, in which the symptoms are slight, perhaps a permanent cessation of the movements can be obtained. In nerve-stretching, which at one time cured almost every disease known to neurologists, but which at the present time has rather fallen into disrepute, we have the means of completely arresting athetosis by producing permanent paralysis of the extremity by severe stretching; or we can produce temporary cessation of the movements, unaccompanied by paralysis, by employing a lesser degree of force. In four cases in which there was marked improvement, the result

was obtained by nerve-stretching in three of them; in the fourth case galvanism is credited with producing the cure.

The first case in which any permanent improvement was obtained, was in a case reported by Dr. Gowers,[1] in 1876. In this case the athetosis followed an attack of hemiplegia; the paralysis was slight, as was also the resulting athetosis, but movements could plainly be observed in both the hand and foot. The patient had never had any convulsions. He was treated with iodide of potash and bromide of potash, and sent on a sea voyage. He returned without showing any improvement. He was then treated with the constant galvanic current. The positive pole was applied to the nape of the neck, and the negative pole rubbed over the overacting muscles. This treatment was continued for two months, with the result that the athetoic movements gradually diminished and finally ceased, in the arm, but no improvement was observed in the leg.

The second case was one reported by Dr. W. J. Morton.[2] The patient was of special interest, as he was the second case on record, and the first one observed in Great Britain. A report of the case was made by Dr. W. T. Gairdner, in the *Journal of Mental Science* for July, 1873. The history of the patient is as follows: When he was three years old he had an attack of hemiplegia, which was not attended by loss of consciousness, nor by convulsions. Three months after the attack, athetoic movements began to show themselves in the hand. The paralysis gradually disappeared, till at the age of eleven he had entirely recovered. The athetosis, in the meanwhile, steadily grew worse. He came to this country and presented himself for treatment at Dr. Hammond's clinic. Dr. Morton took him in charge, and after all medicinal and electrical treatment had failed to give any relief, he decided to stretch the median and ulnar nerves. The operation was performed on November 16, 1882, and was, I believe, the first time that nerve-stretching had been resorted to for the relief of athetosis. While the patient was under the influence of ether all athetoic move-

[1] *Med. Chirurg. Trans.*, London, 1876, vol. lix.

[2] JOURNAL NERV. AND MENTAL DISEASES, 1882, N. S., vii.

ments ceased. The nerves were very forcibly stretched. Quoting from Dr. Morton's report, he says: "Using the nerve as a loop, and the index finger as a crook, the extended arm was lifted again and again from the table, and pulled strongly downwards, so as to pull upon the spinal cord. The ulnar nerve was treated in the same manner. After the patient recovered from the anæsthesia, it was found that the hand and forearm were motionless. The last time Dr. Morton saw the case, between two and three months after the operation, the hand was still paralyzed.

The third case, which I am fortunately able to present to you to-night, is one of great interest, both from a clinical and historical standpoint. He is the original case whose symptoms were first described by Dr. Hammond[1] in 1871. His history is briefly as follows: In 1860 he had an epileptic attack, and has continued having them ever since, sometimes as often as four or five times a week. His habits were bad. He was a gin-drinker, and he states that he has often taken as many as sixty glasses of gin in one day. In 1865 he had an attack of delirium tremens, and remained in a more or less unconscious state for six weeks. After recovering his intelligence he noticed a sensation of numbness in the right arm and leg; severe pain also appeared in these parts, which was shortly followed by the complex movements described under the name of athetosis. At first these movements could be controlled by the will, and ceased entirely during sleep, but gradually, as the athetosis advanced, the will lost its power over the muscles, and eventually the motions were present both day and night. The pain in the arm and leg was intense and frequently kept him awake all night.

I saw him in the spring of 1882. Various medicinal remedies and the different forms of electricity had been used without any beneficial result. I decided to stretch the median nerve, and on May 27, 1882, I performed the operation at the Metropolitan Throat Hospital. While the patient was under the influence of ether no athetoic movements were observed. The median nerve was exposed

[1] "Diseases of Nervous System."

at the inner edge of the biceps, and was apparently in a healthy condition. Passing the finger underneath the nerve, strong traction was brought to bear on it, principally in a downward direction. On the patient's recovery from the ether it was found that the athetosis had entirely ceased both in the arm and in the foot, and that the muscles in both extremities were completely under the control of the will. It was subsequently ascertained that his epileptic paroxysms also ceased, and the pains in both the arm and leg completely disappeared. At the time of the operation he was having from four to six attacks weekly. I exhibited the patient that summer before the American Neurological Association as a case of athetosis cured by nerve-stretching. But in that I was a little premature, as the case will show. About three months after the operation the patient had an epileptic fit. As time went on he had more of them, the pains returned in the arm and foot, and the athetoic movements began slowly to make their appearance, so that in about five months after the operation he was as bad as ever. In 1884 I operated on him again. I exposed the nerve in about the same place. It still appeared healthy. The stretching was performed in the same way as in the first operation, and was attended with the same results. That is, the athetosis disappeared in both hand and foot, the pain ceased, and the convulsions were arrested. He remained well for about four months, then his symptoms reappeared again, and in about eight months' time they were as bad as ever. On February 17, 1885, I operated in the same manner as on the two previous occasions. This time the nerve did not appear to be in such good condition. It had lost its white glistening color and seemed grayish ; it was soft and had lost its elasticity, so that when I stretched it a loop about three inches in length remained outside the wound. The results obtained were the same as in the other two operations. The only difference is that this time they have been more lasting. It is now sixteen months since the operation was performed, and so far as the athetosis goes he is almost recovered. He is able to use his hand in writing, dressing himself, eating, and in fact for

almost any purpose. The muscles of the arm, hand, and foot are perfectly under the control of the will. He can perform extension or flexion, or retain his fingers and toes in any desired position as long as he pleases. When his attention is not directed towards his hand, it is seen to resume its old position, though the motions are absent. This may be the result of habit, as he has been forced to carry his arm in that position for over twenty years. He has an occasional epileptic paroxysm about once or twice a month. He also suffers from that interesting epileptic condition known as double consciousness. He has been for as long a period as four days at a time in that state, apparently perfectly rational, but not being conscious of a single act.

How long his present condition may last, it is of course impossible to state. In view of my past experience in this case, I do not venture for a moment to call this man cured. I would state, however, that I think more improvement has been obtained from nerve-stretching than has ever been secured from the use of electricity or any internal remedies. It will at once occur to you to ask how it is possible, by stretching the median nerve in the arm, to arrest athetoid movements in the foot, and to control epileptic seizures. The only answer I can make to this question is to say that I don't know. That he had the symptoms previously referred to, is a matter of history about which there can be no doubt, and that he has not them now is self-evident. No one would pretend for a moment that such organic lesions as are known to produce athetosis could be removed by stretching a nerve in the arm. If the man had only had athetoic movements in the arm, or if only the movements in the arm had ceased, the improvement could very readily be attributed to a local effect upon the nerve at the point where it was stretched. But the complete cessation of the movements in the hand and foot, the relief of the epileptic seizures, and the intense pain, and the return of the muscles to the control of the will, would seem to indicate that stretching of the nerve has produced such an effect upon the diseased organs that the generation of athetoic impulses becomes impossible. This hypothesis is not so improbable

as it would at first seem to be. Dr. Dana showed conclusively that stretching the sciatic nerve also stretched the spinal cord as high up as the cervical region, and perhaps higher. It therefore does not require that the imagination should be stretched in order to comprehend how stretching the median nerve, which is so much higher up than the sciatic, could influence the corpus striatum or even the cortex. That nerve-stretching holds out the only hope of relief for athetosis, I am firmly convinced. An interesting case is reported by Griedenberg.[1] One of his cases of athetosis was operated on by Dr. Fricke, on August 24, 1882. "The median nerve was stretched, using the finger as a hook. Immediately after the operation, and on the following day, no movements were noticed. On the second day after the operation the athetosis re-appeared, and on the fourth day the movements had regained their former intensity. The disease then remained *in statu quo ante*." It is quite likely that subsequent operations would have been attended with longer intervals of rest.

There is undoubtedly a close connection between all forms of muscular spasm and incoördination due to cerebral disease, as exhibited variously in tremor, clonic spasm, chorea, ataxia, and athetosis; and some authors claim that a lesion in any part of the motor tract, from the cortex cerebri to the spinal cord, may give rise to any of these forms of motor spasms. That this is true of tonic spasm, is proved beyond a doubt by the enormous number of cases with which you are all familiar, and I think it may be shown by the same cases that rigidity and tonic contractions are due to irritation of nerve fibres and not of nerve cells. On the other hand, mobile spasms, as exhibited by chorea, ataxia, and athetosis, depend upon irritation of motor or coördinating cells. There is no evidence to show that a lesion only affecting the motor fibres in the brain has ever been followed by mobile spasms of any kind. On the contrary, the large number of autopsies held on persons who have died while suffering from cerebral chorea, ataxia, and athetosis, invariably present lesions of the cortex or basal

[1] "Vier Falle von Athetose." *St. Petersburg Med. Wochenschr.*, 1882, vii.

ganglia, and when rigidity and tonic spasms are also present the motor fibres are found to be involved in the lesion. In an interesting paper on this subject, Dr. Emil Denange[1] reports eleven cases of cerebral ataxia, chorea, and athetosis, accompanied in many cases by hemiplegia and hemianæsthesia. In some of the cases the disease appeared subsequently on the other side of the body. In nine of these cases autopsies were obtained, and in every one, with the exception of the case of athetosis, a lesion was found either in the corpus striatum, and particularly in the lenticular nucleus, or else the optic thalami were involved. In those cases in which the disease became bilateral, a similar lesion, usually a patch of softening, was discovered in the lenticular nucleus or optic thalamus upon the opposite side. When there was rigidity and hemiplegia, the lesion was observed to affect the motor nerve fibres in the internal capsule.

In the case of athetosis the lesion was found to consist of extensive softening of the cortex-cerebri in the region occupied by the motor centres. Many other similar cases are reported by Charcot, Weir Mitchell, and others.

Dr. S. J. Sharkey,[2] in his exhaustive lecture on muscular spasms, details a large number of cases of tonic spasms and two cases of athetosis. In those cases in which the cortex was involved, tonic spasms followed the paralysis and the post-mortem disclosed degeneration of the motor fibres. In none of the other cases of tonic spasm were the basal ganglia involved, except in one case where there was a tubercular mass in both optic thalami, but in this case there was also tremor. In the two cases of athetosis no post-mortem was held, as the patients did not die. Dr. Sharkey also reports one case of almost complete destruction of one corpus striatum, in which no mobile spasm existed, the patient merely suffering from a temporary hemiplegia, and quotes this case in support of the theory that athetosis and the allied disorders do not depend upon disease of this organ for their existence. But I do not think this one case proves that his views are correct. If mobile

[1] *Revue de Médicine*, May, 1883.

[2] Gulstonian Lectures, *Brit. Med. Journ.*, March 27, 1886.

spasm depends upon irritation of the cells in the corpus striatum, the cortex, and of the optic thalamus as well, the destruction of these organs would undoubtedly prevent the conception of spasmodic impulses. It is only when the destruction is partial, when the cells of these organs are not destroyed, but only irritated, that mobile spasms become possible.

Nothnagel cites several cases of lesion of the lenticular nucleus, followed by hemiplegia, but unaccompanied by mobile spasms, but his cases all died within ten days after the lesion occurred, and before they had time to get athetosis or any thing else.

I think that a careful study of these cases will show pretty conclusively that cerebral mobile spasms, such as tremors, ataxia, chorea, and athetosis, depend upon lesions of the corpus striatum, the optic thalamus, and the cortex cerebri, and not upon disease of the motor nerve fibres, nor any other part of the motor tract.

A lesion simply involving the corpus striatum or optic thalamus does not necessarily produce hemiplegia nor hemianæsthesia. The motor fibres do not pass through the corpus striatum at all, but if a hemorrhage occurring in the corpus striatum is extensive enough, it will exert pressure upon the motor fibres as they pass through the internal capsule, and thus produce a hemiplegia which will gradually disappear as the absorption of the hemorrhage takes place. This is what occurs in a large proportion of the cases of post-hemiplegic athetosis. If the lesion in the corpus striatum does not interfere with the internal capsule, athetosis may result without hemiplegia. In those cases in which the hemiplegia becomes permanent there is undoubted rupture of the motor fibres, but the corpus striatum or the cortex must be included in the lesion as well. I do not think there is a single case on record of post-hemiplegic athetosis in which the lesion was found to be confined entirely to the motor nerve fibres. I have been able to secure the histories of nine cases on whom post-mortem examinations were held. The first case, according to Brissaud,[1] was reported by Lauenstein. A detailed history of the

[1] *Gaz. Hebd.*, 1880, p. 803.

case is not given, but the autopsy disclosed a lesion involving the posterior part of one optic thalamus. The corpora striata and cortex were healthy.

The second case was reported by Pick.[1] Here also the lesion was found to exist in the posterior portion of the optic thalamus.

Grasset[2] details the result of an autopsy held on a case of athetosis. In this case there were three spots of softening, one on the inferior portion of the optic thalamus, one in a portion of the caudate nucleus, and one in the lenticular nucleus.

The fourth case was one of Richets, but was reported by Oulmont. In this case there were several spots of softening in different parts of the hemispheres. There was also an area of softening which destroyed almost the entire posterior portion of the caudate nucleus, and another area which had made a deep cavity in the lenticular nucleus, which further on crossed the internal capsule and joined the lesion in the caudate nucleus.

The fifth case came under the observation of Dr. Fletcher Beach.[3] There had never been any hemiplegia, but the patient had suffered for some years with epileptic seizures. The necropsy was held thirty-six hours after death. The corpus striatum and optic thalamus appeared to be healthy. A microscopical examination was made of the rest of the brain substance, but a similar examination of the corpus striatum and optic thalamus was, for some reason, not deemed necessary. The microscope revealed an increase in number of the vessels, distension of many of them, extensive infiltration of the tissue with leucocytes, especially in the perivascular sheaths of the vessels, and many of the vessels contained clots. These changes were principally in the cortex of the inferior parietal lobule, and first temporo-sphenoidal convolution. It is very likely that the microscope might have demonstrated similar changes in the corpus striatum and optic thalamus.

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[1] *Prager Vierteljahrschrift*, 1879, p. 141.

[2] *Prog. Méd.*, Paris, Nov. 13, 1880.

[3] "An Account of the Microscopical Appearances in a Case of Athetosis." *Brit. Med. Journal*, 1880, i., 967.

In the sixth case, reported by Ringer,[1] the athetosis had been preceded by hemiplegia, hemianæsthesia, and aphasia. Before the athetosis appeared he had regained his speech, the anæsthesia disappeared and the hemiplegia was partially recovered from. The autopsy showed the left optic thalamus to be smaller and flatter than the right one. There was a cyst occupying the posterior part of the lenticular nucleus, and involving the white matter outside and beneath the thalamus and a small part of the thalamus itself. About one fifth of the lenticular nucleus was destroyed, together with a few fibres of the internal capsule.

The seventh case was reported by Landouzey.[2] There was nothing apparently in this case to account for the disease. The patient was about thirty-two years of age, and had had athetosis from childhood. She had never suffered from epilepsy, nor had she ever had hemiplegia. The autopsy revealed a focus of softening in the extra-ventricular nucleus of the corpus striatum, on the left side. In the centre of this patch of softening, a calculus about the size of a bean, was found. The athetoid movements were in both the hand and foot, on the right side.

The eighth case was reported by Dr. Murrell.[3] The patient at the time of death was thirty-three years of age. When he was three years old he had an attack of whooping cough, and soon afterwards two epileptic attacks, which left him paralyzed down the left side. The paralysis gradually disappeared, so that when he was ten years of age he could run and play like other boys. As the paralysis left him, athetoid movements began to show themselves in both arm and leg, but were always very slight in the leg. The patient died from phthisis. The necropsy was held thirty-six hours after death. The whole right hemisphere was smaller and about three quarters of an inch shorter than the left one. Almost the entire lenticular nucleus was destroyed. The posterior part of the caudate nucleus was unaffected.

[1] "Notes of a Post-Mortem Examination in a Case of Athetosis." *Practitioner*, London, Sept., 1879.

[2] *Progrès Méd.*, 1878, Nos. 5 and 6.

[3] "Case of Athetosis. Death from Phthisis. Post-Mortem." *Lancet*, Lond., 1879, i., 369.

The ninth case was reported by Emil Denange.[1] The patient had suffered from hemiplegia, which was followed by athetosis in the hand. The autopsy showed that the corpus striatum and optic thalamus were perfectly healthy. There was a large patch of softening on the cortex, which involved all that portion of the posterior ascending convolution in which Ferrier locates centres for the complex movements of the finger and hand.

It is therefore evident, from a study of these cases, that in three out of the nine the lesion was found to be in the corpus striatum; in two others in the optic thalamus; in three others both the corpus striatum and optic thalamus were involved; while in the other two cases a diseased condition of the cortex existed.

In regard to the situation of the lesions in chorea, ataxia, and tremor, they may be said to be identical with those of athetosis.

Gowers [2] reports a case of a patient who had an attack of hemiplegia, which came on somewhat deliberately in the course of an hour, was never quite absolute, and soon lessened. The ataxy developed with the recovery of power. It was present in the arm only, and was not accompanied by spontaneous mobile spasms. The incoördination was at one time so violent that if the patient raised an object from the table the arm would fly up over the head. The autopsy revealed a mass of cicatricial tissue through the middle of the optic thalamus, and was evidently the remains of an old focus of softening. The corpus striatum was not affected.

Denange [3] reports nine cases embracing tremor, chorea, and ataxia, in which one or both of the basal ganglia were involved. In one case of ataxia the disease was found to be due to a cortical lesion. In every case where the internal capsule was included in the lesion rigid spasm was also present.

Weir Mitchell reports two cases of post-hemiplegic ataxia. In both cases the lesion was found in the optic thalamus.

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[1] *Revue de Médecine*, Paris, May, 1883.

[2] *Med.-Chirurg. Review*, London, 1876, vol. lix.

[3] *Op. cit.*

Sharkey [1] reports two cases, one of tremor accompanied by paralysis and rigidity, in which both thalami were invaded by tubercular deposits, which also pressed upon the internal capsule; the other was a case of ataxia, in which the lesion was found to exist in the cortex.

It is mainly from a study of autopsies that we are able to derive any definite knowledge of diseases of this character, and if autopsies prove any thing, I think these show that athetosis and its allied disorders depend upon lesions of the corpus striatum, optic thalamus, and cortex cerebri, and of these parts only.

[1] *Op. cit.*